




Assessment of Functional Outcome Following Duhamel Retro-Rectal Pull-Through Surgery for Hirschsprung's Disease - A Follow-up Study

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J Coloproctol 2023;43(4):e271–e275.

Abstract

Context Hirschsprung's disease (HD) is one of the commonest problems requiring surgery in children. More than 95% of children present during new-born period, when they are treated with leveling colostomy and are followed with pull-through surgery a few months later, once the child has gained adequate weight to withstand a major surgery. The commonest pull through surgery done is the Duhamel retro-rectal pull-through (DRPT) repair.

Settings and Design This is a retrospective study of children who presented to one unit in our institute, a tertiary care referral hospital for children less than 12 years, with HD and underwent DRPT procedure during the period between July 2017 to June 2020. The children were evaluated after three years of follow-up for fecal incontinence and constipation. The study was conducted in children diagnosed with classical segment recto-sigmoid HD who underwent surgery. The children who were diagnosed with HD other than classical segment, who underwent primary pull through surgery and who underwent other repairs for HD were excluded from the study.

Results Thirty-two children underwent DRPT procedure during the study period. Of them, five (15.6%) children were lost on follow-up and one (3.1%) child had expired in the immediate post-operative period. Twenty-six children were included in the study. The bowel function score was calculated. The mean age of definitive surgery was 4.2 years. The follow-up period was a minimum of three years. Only two children had a "good" score of eighteen and above. Nineteen children had a "fair" score of 13–17. Five children had a "poor" score of less than thirteen, and among them, two had a "very poor" score of less than nine. The mean BFS was 13.72.

Conclusions Functional outcomes following Duhamel procedure are satisfactory, with 7.7% of children are in the fringe of requiring another surgery for constipation and pseudo-incontinence.

Keywords

- ▶ Hirschsprung's disease
- ▶ Duhamel procedure
- ▶ bowel function score
- ▶ constipation
- ▶ pseudo-incontinence
- ▶ MACE - Malone's antegrade colonic enemas

received
August 8, 2023
accepted after revision
October 24, 2023

DOI <https://doi.org/10.1055/s-0043-1776891>.
ISSN 2237-9363.

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Thieme Revinter Publicações Ltda., Rua do Matoso 170, Rio de Janeiro, RJ, CEP 20270-135, Brazil

Key Messages

HD is one of the commonest presentations in the newborn period. Duhamel procedure is one of the commonest definitive surgical procedures for HD, as it is easily reproducible, having lesser complications. Assessment of functional outcome following definitive procedure is an important point in the follow-up period.

Introduction

HD is a common cause of intestinal obstruction in newborn, requiring surgery. It presents with functional obstruction caused by the aganglionosis of the colon, which is due to the failure of the migration of the neural crest cells in the bowel,^{1,2} mainly the distal bowel, beginning at the internal sphincter and extending proximally for varying distances. The incidence of HD ranges from 1 in 5000 to 1 in 12000 live births.³ It has a male preponderance with a male: female ratio of 4:1.

Classical HD is routinely treated with leveling colostomy at newborn and a definitive repair⁴ is done once the child has attained adequate weight. Among the several pull-through surgical repairs, DRPT repair is one of the commonest, as it is easily reproducible and has limited complications. Most of the studies have mainly included children who have been operated using the DRPT procedure for the recto-sigmoid HD.⁵ But nowadays, primary transanal endo-rectal pull-through is being preferred.⁶ The functional outcome of the pull-through surgery needs to be assessed post-operatively in a long-term follow-up period.⁷ Functional outcome is defined by the child's ability to control defecation, social function and in the long term, by the quality of life.⁸

Subjects and Methods

This is a retrospective study of children who presented to one unit in our institute, a tertiary care referral hospital for children less than 12 years, with classical segment HD and underwent DRPT repair during the period between July 2017 to June 2020 and had completed at least three years of follow-up.

Inclusion criteria were classical segment HD children who underwent DRPT repair, after having been diagnosed and undergoing leveling colostomy at the time of diagnosis and are on follow-up for a period of at least three years. The children who were diagnosed with HD other than classical segment, who underwent primary pull through procedures, who underwent other pull-through repairs and who were lost on follow-up or were expired after surgery were excluded from the study. Inclusion and exclusion criteria are summarized in ► **Figure 1**.

Children data were collected from the case sheets and the following were obtained: age, gender, symptoms, clinical diagnosis, radiological imaging, medical management given, surgery performed, post-operative complications, any other surgical procedure needed for the post-operative complications, post-operative histo-pathological report, follow-up and functional outcome.

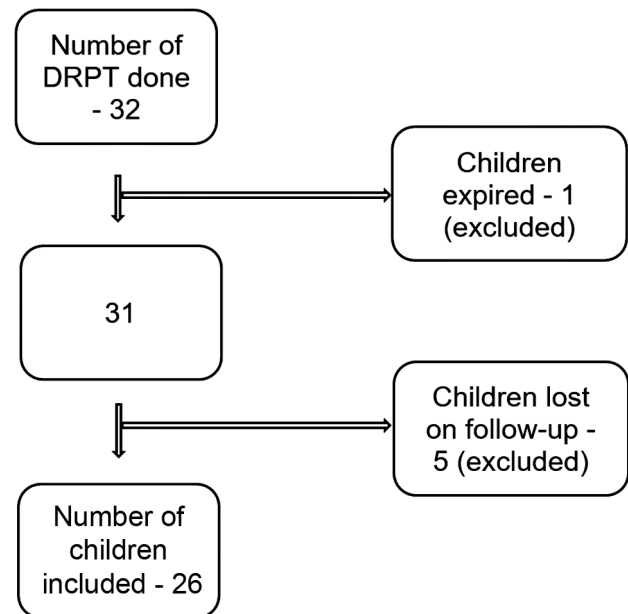


Fig. 1 Flowchart of study inclusion and exclusion criteria.

As part of a routine, all children were evaluated pre-operatively and were posted for surgery only if they had normal hemoglobin and albumin levels. All children underwent modified Duhamel pull-through procedure, with closure of the stump well below the peritoneal reflection, coloanal anastomosis at ~0.5 cm above the dentate line posteriorly and usage of linear cutting stapler between the posterior wall of native rectum and the anterior wall of pull through bowel. After surgery, all children were started on clear liquids once the ileus settled and passed stools. All children were discharged on the seventh post-operative day, if no complications occurred, after they were started on and tolerated oral feeds. The histo-pathological report of the doughnut specimen of the pull-through bowel was obtained before the day of discharge and checked for the presence of ganglion cells. All children were followed up on the 14th day, when sutures were removed, and a digital rectal examination was done to assess for the residual spur. The children were followed up once a month initially for three months and then every three months, thereafter.

Functional outcome was assessed using Rintala's bowel function score (BFS),⁹ at the time of follow-up after a period of three years. Some children needed admission for the management of their bowel problems during the follow-up period. The score was calculated by interviewing the parents with regard to the following seven variables: ability to hold back defecations, feels/ report the urge to defecate, frequency of defecation, soiling, fecal accidents, constipation and social problems. Each variable is scored from 0–3, except for the frequency of defecation, which is scored 1–2. The maximum score amounts to 20. Soiling is defined as the staining of the underwear or involuntary loss of small amounts of stool.^{9,10} Accidents are defined as the involuntary loss of large amounts of stool requiring change of the underwear. Soiling and accidents denote fecal incontinence. The score is

classified as good, if it is 18 or more, fair, if it is between 13–17, poor, if it is below 13 and very poor, if it is below nine.¹¹

If any children needed admission for their bowel problems, they were scored using BFS and they were divided into constipated or incontinent group. Accordingly, they were evaluated if they didn't get cured with bowel management program. The constipated group underwent repeat bowel enema to assess for any residual stricture or twist of the pull-through bowel segment, repeat biopsy to look for residual or acquired aganglionosis of the pull-through bowel segment and anorectal manometry to look for the presence of recto-anal inhibitory reflex.

The incontinent group were evaluated for real incontinence or pseudo-incontinence (overflow) and were treated accordingly.

Data were analyzed. Quantitative data were described as numbers and percentages. Proper ethical clearance was obtained from the institution ethical committee. Proper informed consent was obtained from the parent of the children for using their data.

Results

Thirty-two children underwent DRPT procedure during the study period. Twenty-seven (84.4%) children were diagnosed with HD in the newborn/ early infancy period and underwent levelling colostomy then. The remaining five (15.6%) children were diagnosed in a later age group between 5 - 11 years. Among the thirty-two children, five children were lost on follow-up. One child expired in the immediate post-operative period, with stump leak and associated peritonitis and re-laparotomy complications. Twenty-six children were included in the study. Among them, seventeen were boys and the remaining nine were girls. The median age of children in the study who underwent DRPT repair was one (range 6 months to 12 years), with a mean of 4.2 years. Clinical characteristics and the demographics of the study population are given in ►Table 1.

Among these twenty-six children, six (23.1%) required a second surgery, either in the immediate post-operative period or in the follow-up period. Among them, four children required had to undergo re-laparotomy in the immediate post-operative period due to stump leak in three children (11.5%) and incidental malrotation in one child, who presented with bilious vomiting in the immediate post-operative period. Remaining two children needed laparotomy due to adhesive small bowel obstruction. Post-operative events and their incidences are summarized in ►Table 2.

The three children who needed re-laparotomy for stump leak underwent stump closure with a covering ileostomy. They were followed up and underwent ileostomy closure after a period of six months, after evaluating with a distal stoma water soluble contrast study and a contrast enema to rule out stricture or stump leak. These children were also followed up after a period of three years from their last surgery (ileostomy closure). The children who presented with adhesive intestinal obstruction needed only adhesiolysis.

Table 1 Demographics of the study children

S. No.	PARAMETER	NUMBER (%)
1	Total children who underwent DRPT	32
2	HD diagnosed at newborn	27 (84.4%)
3	Late diagnosis of HD	5 (15.6%)
4	Total participants	26
5	Boys	17 (65.4%)
6	Girls	9 (34.6%)
7	Mean age of DRPT	2.86 years
8	Mean duration of follow-up	4.2 years

Abbreviation: DRPT, Duhamel retro-rectal pull through; HD, Hirschsprung's disease.

Table 2 Post-operative events

S. No.	INTERVENTION	NUMBER OF CHILDREN (%)
1	Need for re-laparotomy	6 (23.1%)
2	Need for ileostomy	3 (11.5%)
3	Mortality	1 (3.1%)
4	Disimpaction of stools	5 (19.2%)
5	Retrograde colonic saline washouts	5 (19.2%)
6	Bowel management program	5 (19.2%)
7	Rectal biopsy	0
8	Anorectal manometry	3 (11.5%)
9	Twisting of pull through bowel	1 (3.8%)
10	Stricture of pull-through segment	0
10	Need for revision pull-through	0
11	Need for rectal spur division	0
12	Need for MACEs	2 (7.7%)

Abbreviation: MACEs, Malone's antegrade colonic enemas.

Bowel Function Score

The minimum follow-up period was three years in the study. Two children had a "good" BFS score of 18 or more. Nineteen children had a "fair" BFS score of 13–17. Five children had a "poor" BFS score of less than 13, with two among them had a "very poor" BFS of less than nine.

None of the children had a "perfect score" of 20 in BFS. Only two children had a "good" BFS score of more than or equal to 18. They had an occasional staining episode, probably less than a week and constipation which was managed by diet alone.

Nineteen children had a "fair" score of 13–17. These children had occasional problem to hold back defecation

and feels the urge to defecation on most of the times and not always. They had occasional soiling, on less than a week and their constipation was managed either by diet or laxatives. Most of them needed laxatives as part of their bowel management program.

Five of the remaining children had a score of less than 13. Among them, three children had a “poor” score between 9–13. They had frequent staining of the underwear and need to change the underwear often. They also had severe constipation which was managed often by saline enemas. The remaining two children had a “very poor” score of less than 9. Among those two children, one had an extremely rare association of the diagnosis of HD after the child was treated for intermediate ano-rectal malformation in three stages. The child had a BFS of 6 and developed incontinence having accidents, requiring protective aids both in the day and night. This child was diagnosed as HD after the stage III procedure for ARM (i.e.) colostomy closure, when she developed abdominal distension, following she underwent diversion ileostomy and then re-evaluated for HD. After confirmation of HD, she underwent Duhamel retrorectal pull-through procedure. The pull-through was difficult as it wasn't a virgin plane, as creation of retro-rectal space was difficult. The other child had severe constipation with frequent pseudo-incontinence (overflow), with a BFS score of 8.

Discussion

In our study, majority of the children had a diagnosis of HD at newborn. Twenty-seven out of thirty-two children were diagnosed at newborn or in the early infant period and underwent levelling colostomy at that time itself. Only five children were diagnosed at a later age (after one year of life). In our study, the male to female ratio was less than 1.5:1, and not the typical 4:1 as seen commonly or in most of the studies. None of the children had Trisomy 21 association.

Twenty children had the definitive repair prior to 1.5 years of age and twenty-three children had the definitive procedure prior to three years of age. Five children underwent DRPT repair after their five years of age. But these children had a different set of difficulties. The problems were encountered during the creation of levelling colostomy, as these children had severe dilatation of the rectosigmoid and the proximal bowel as well. The dilated distal bowel segment has to be excised during the procedure and divided colostomy needed to be done. Postoperatively, there was also stomal prolapse in two children, which needed to be reduced prior to the pull-through procedure.

Two children among them had their repairs done beyond ten years of age. There was no increase in complications between those children who were operated for definitive repair in early age than in children who underwent repairs in their later ages. There was one mortality (3.1%) in those who had undergone the pull through repair during our study period. The child had a massive stump leak and expired due to the complications of leak and the required surgery.

Five children (19.2%) needed admission for their stooling complications. These children needed disimpaction of hard

stools and retrograde saline washouts as part of their bowel management program. Three children underwent anorectal manometry as part of their evaluation for persistent symptoms and it was not contributory. None of the children needed rectal biopsy. One child was diagnosed to have twisting of the pull-through bowel based on the contrast enema.

Comparing the BFS between girls and boys, girls fared marginally better than the boys (mean BFS - 14.9 in boys versus 16 in girls). The prevalence of constipation at some point of time in the study children (38.5%) compares similar to other series after DRPT.¹² The prevalence of constipation at any point of time was less in girls when compared with boys (7 in boys versus 3 in girls - 41.2% in boys versus 33.35% in girls). After correction of constipation with stool softeners and enemas, these prevalence rates came down. Only five children needed evaluation for their persistent constipation shows that the constipation prevalence rates came down (19.2%) at the end of three years after surgery.¹³

Frequency of stooling pattern changed from the initial days of surgery, when they had stool frequencies close to ten times a day to ~2–3 stool frequency per day at about one year after surgery.^{14,15} Eight out of twenty-six children (30.8%) had soiling at the end of their three years after surgery, which is lower when compared with other studies (48%).

None of the children had enterocolitis like symptoms after the surgery. None of the children had stricture of the pull through bowel. No children had a long residual spur as a cause for their constipation. No children needed re-do pull through procedure as there was no persistent/ acquired aganglionosis in this group of children and hence none of the children even needed rectal biopsy for their evaluation of persistent constipation.

When BFS more than or equal to 17 is taken as the upper limit of normal or “satisfactory” BFS,¹⁶ as in a series by Rintala and others, 61.5% had a normal bowel function, when compared with only 47% in the compared series. Social problems related to bowel function persisted in 19.2% in our study group which is lesser when compared with the Rintala study group (29%).

Intractable symptoms are persisting in five children and they are managed by bowel management program. Two children (7.7%) with “very poor” BFS are in the verge of needing another procedure in the form of Malone's antegrade colonic enemas, when compared with 3% in Heman-shoo S. Thakkar et al study.

Quality of Life (QOL)

The assessment of QOL is a much more difficult question. In general, QOL issues have been poorly described in most reviews. According to Moore et al., “*quality of life remains a difficult concept to assess and is influenced by the physical, psychological, spiritual, functional and social well-being of the individual.... Functional results are central to quality of life.*”

Overall QOL is described as quite good, as only five children (19.2%) had intractable symptoms and even among them, three children were managed efficiently by the bowel

management program and became well-adjusted children. Only two children (7.7%) were refractory to the bowel management program. Hence, the remaining children (92.3%) had a good QOL, which is comparable to other series.

Conclusion

As with most operations, complications arising even years after surgery can be attributed to problems occurring during the operation itself. Most children had an excellent outcome after their DRPT procedure. Among the five children with a “poor” BFS, three children were well managed with a conservative, non-operative approach with a successful outcome. In only two children, problems continue to persist and needs another surgery for their bowel management.

According to Orvar Swenson, “*Resection of the aganglionic colon.... Is a difficult operation. Yet, if a well-trained surgeon has an opportunity to observe the technical details of the operation and then perseveres, good results can be obtained.*” However, even after the best operation, problems may persist following the definitive repair for HD.

Conflict of Interest

None.

Acknowledgment

Great thanks and warmest gratitude to the patients, their caretakers and to all department members in our hospital who helped to treat the children and to all who provided us with all we needed to work on the study cases effectively.

References

- 1 Gunadi A, Monica Carissa T, Stevie A, et al. Long-term functional outcomes of patients with Hirschsprung disease following pull-through. *BMC Pediatr* 2022;22(01):246
- 2 Puri P, Nakamura H. Epidemiology and clinical characteristics of Hirschsprung's disease. In: Puri P, ed. *Hirschsprung's Disease and Allied Disorders*. Cham: Springer International Publishing; 2019: 167–172
- 3 Tilghman JM, Ling AY, Turner TN, et al. Molecular genetic anatomy and risk profile of Hirschsprung's disease. *N Engl J Med* 2019;380(15):1421–1432
- 4 Taguchi T, Matsufuji H, Ieiri S, et al. Hirschsprung's Disease and the Allied Disorders. In: *Status Quo and Future Prospects of Treatment*. 1st ed. Singapore: Springer Singapore; 2019. Doi: 10.1007/978-981-13-3606-5
- 5 Conway SJ, Craigie RJ, Cooper LH, et al. Early adult outcome of the Duhamel procedure for left-sided Hirschsprung disease—a prospective serial assessment study. *J Pediatr Surg* 2007;42(08): 1429–1432
- 6 Huang EY, Tolley EA, Blakely ML, Langham MR. Changes in hospital utilization and management of Hirschsprung disease: analysis using the kids' inpatient database. *Ann Surg* 2013;257(02):371–375
- 7 Nasr A, Grandpierre V, Sullivan KJ, Wong CA, Benchimol EI. Long-term outcomes of patients surgically treated for Hirschsprung disease. *J Can Assoc Gastroenterol* 2020;4(05):201–206
- 8 Drissi F, Meurette G, Baayen C, et al. Long-term outcome of Hirschsprung disease: impact on quality of life and social condition at adult age. *Dis Colon Rectum* 2019;62(06): 727–732
- 9 Rintala RJ, Lindahl HG, Rasanen M. Do children with repaired low anorectal malformations have normal bowel function? *J Pediatr Surg* 1997;32(06):823–826
- 10 van der Steeg HJJ, van Rooij IALM, Iacobelli BD, et al. On-behalf-of-the-ARM-Net-Consortium. Bowel function and associated risk factors at preschool and early childhood age in children with anorectal malformation type rectovestibular fistula: An ARM-Net consortium study. *J Pediatr Surg* 2022;57(09):89–96
- 11 Ure BM, Rintala RJ, Holschneider AM. Scoring postoperative results. In: Holschneider AM, Hutson JM, eds. *Anorectal Malformations in Children*. New York: Springer, Berlin Heidelberg; 2006:351–359
- 12 Jarvi K, Laitakari EM, Koivusalo A, Rintala RJ, Pakarinen MP. Bowel function and gastrointestinal quality of life among adults operated for Hirschsprung disease during childhood: a population-based study. *Ann Surg* 2010;252(06):977–981
- 13 Heikkinen M, Rintala RJ, Louhimo I. Bowel function and quality of life in adult patients with operated Hirschsprung's disease. *Pediatr Surg Int* 1995;10:342–344
- 14 Davidson JR, Kyrklund K, Eaton S, et al. Long-term surgical and patient-reported outcomes of Hirschsprung Disease. *J Pediatr Surg* 2021;56(09):1502–1511
- 15 Lawal TA, Chatoorgoon K, Collins MH, Coe A, Peña A, Levitt MA. Redo pull-through in Hirschsprung's [corrected] disease for obstructive symptoms due to residual aganglionosis and transition zone bowel. *J Pediatr Surg* 2011;46(02):342–347
- 16 Nah SA, de Coppi P, Kiely EM, et al. Duhamel pull-through for Hirschsprung disease: a comparison of open and laparoscopic techniques. *J Pediatr Surg* 2012;47(02):308–312